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Carcinoid Tumors of the Colon and Rectum

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IN 1808 MERLING first cited the carcinoid as an appendiceal tumor.[1] Five to ten percent of all carcinoid tumors are located in the colon, with the majority occurring in the cecum.[2,3] This tumor is the most common neoplasm of the appendix and is most often found in this organ.[2,4,5] The tumor is present in approximately 0.3 percent[2] of all appendices, variations depending upon the method of reporting. The literature is not clear on carcinoid tumors of "the colon" as reports are related to the appendix, rectum and colon or the colon exclusive of the rectum.[6-9]

Age and Sex

As with other types of tumors, the incidence of carcinoid is highest in middle and old age. Appendicular carcinoid tumors usually occur at 25 to 30 years.[10] The average age of all patients with rectal carcinoids is 55 years.[7] Thirty-nine years is the mean age of patients with malignant rectal carcinoids.[10]

Considering all carcinoid tumors, there is no apparent difference in the incidence in males and females.[10] However, the incidence of carcinoids of the appendix alone is approximately twice as great in females as in males.[10]

Symptoms

Carcinoids of the appendix are usually incidental findings, but occasionally they cause obstruction and are the direct cause of appendicitis. Carcinoids of the colon tend to be large, and the symptoms are similar to those of carcinoma except that carcinoids have less tendency to ulcerate and bleed.[4] Colonic carcinoids tend to be large, the size correlating well with the high rate of metastasis.[9] Most rectal carcinoids produce few symptoms.[6-8,11] Rectal bleeding, change in bowel habits, varying degrees of obstruction, or signs and symptoms of metastatic carcinoma may be present.

Pathology

A yellow character is often noted on cut section. The tumors are usually submucosal. Ulceration is not common, and necrosis rare.[12]

In tumors of the mid-gut, cytoplasmic granules can be demonstrated by the argentaffin reaction.[13] In lesions of the fore-gut and hind-gut, however, silver staining is variable.[13]

The incidence of metastasis from appendiceal lesions is only 3 percent because so often the appendix is removed routinely before dissemination can occur,[4] but from carcinoid tumors of the cecum and colon it is high.[2,4,9] Size is the best criterion by which to predict the malignant potential of these tumors in the rectum.[6,7] Lesions less than 1 cm in diameter are almost invariably benign, and those larger than 2 cm almost always develop local extension or metastasis. In addition to size, invasion into or beyond the true muscularis is an indication of malignancy. For that reason it is imperative that biopsy specimens be deep enough so that invasion can be seen.[6,7]

The reported incidence of multiple carcinoids of the colon[9] and rectum[7] has varied from 2 to 4.5 percent. For other malignant neoplasms associated with carcinoid tumors of the colon[9] and rectum[7] the range is from 2.5 to 38 percent.[25]

Carcinoid Syndrome

Association of the carcinoid syndrome with appendiceal or colonic carcinoids is unusual.[2] There has been only one report of this syndrome in conjunction with a rectal lesion.[14] Ahlgren et al in 1962 published a case of carcinoid tumor of the rectum with moderate rise in serotonin without association of the carcinoid syndrome, the only case of a secreting carcinoid of the rectum described in the literature.[15] Orloff[7] accepts neither of those reports and presents his case well. Ingestion of serotonin-containing foods, such as bananas, tomatoes, avocados, red plums, walnuts and eggplant,[16] and drugs such as methocarbamol and mephensin carbamate,[17] elevate the urinary 5-hydroxyindoleacetic acid (5-HIAA) levels.

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Treatment

The treatment of carcinoid tumors is primarily surgical. When lymphatic permeation[18-20] or tumor at the surgical margin[21] is present, right hemicolectomy is advocated for appendiceal carcinoids. Considering that the prognosis of carcinoids of the appendix is excellent, with only a few deaths reported as directly attributable to the tumor,[22] one must be suspect of retreatment of this lesion regardless of the pathologic findings if the tumor is less than 2 cm in diameter.

In a review of the literature, Berardi[9] noted that the procedure most frequently used for colonic carcinoids was hemicolectomy. Anterior resection is the treatment of choice for sigmoid carcinoids.[9] When a nonresectable tumor was found, palliative colostomy or ileocolostomy was done.[9] In patients with the carcinoid syndrome (widespread metastatic disease) palliative resections to decrease the mass of tumor significantly improved the condition of some patients and prolonged life.[9] As was previously noted, recurrent carcinoids are extremely rare.[23]

Rectal carcinoids less than 2 cm in diameter, without muscle invasion, can be treated by local excision, excisional removal, and fulguration of the bed.[6-8] Larger tumors and those invading the muscle require radical extirpation, usually consisting of an abdominoperineal resection with permanent colostomy.[6-8]

Radiotherapy has proved to be of little value in the treatment of carcinoids except in an occasional case of metastasis to the liver.[24]

Prognosis

The prognosis of carcinoid of the appendix is excellent. Moertel and coworkers[5] reported that in a series of 108 patients traced for more than five years and 83 for more than ten years, there were no recurrences regardless of the degree of local invasion. The same observers reviewed more than a thousand cases in the literature and were able to find only two instances of recurrence.

Approximately one-fourth of all patients with malignant rectal carcinoids die postoperatively or from disease within five years.[7]

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